

Dysplasia epiphysealis hemimelica of the ankle in children

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Dysplasia epiphysealis hemimelica (DEH), also known as Trevor's disease, is a rare developmental disorder affecting the epiphyses; in young children, it usually involves the knee and ankle joints. DEH was first reported by Mouchet and Belot in 1926,¹ who named it tarso megalia. In 1950, Trevor² used the term tarso-epiphyseal aclasis. Fairbank³ in 1956 coined the term dysplasia epiphyseal hemimelica, the term that remains the common usage name for this unusual condition.

The etiology of DEH is unknown. Some authors have theorized that the defect is in the regulation of cartilage proliferation⁴ or that the disorder may be due to a localized disturbance of the pre-

postaxial part of the apical cap of the limb bud in early fetal development.⁵ It does not appear to be genetically transmitted,⁵⁻⁸ and no case of malignant transformation has been reported.⁶⁻¹⁰ The incidence of DEH has been estimated at 1 in 1 000 000.⁸ It is more common in males (3:1) and white Caucasians, and usually presents in childhood or early adolescence. The most common sites affected are around the knee, usually on the medial side of the tibial or femoral epiphysis, and the ankle, particularly the talus and the medial aspect of the fibular and tibial physis (Fig. 1). The natural history of DEH is that of continual growth of the lesion until skeletal maturity, at which time further enlargement ceases.

The aim of this study was to examine the effects of DEH on ankle function and development and to recommend a treatment regime for this unusual but disabling condition.

Methods

Four patients with painful ankles, limited function and deformity secondary to DEH have been assessed at the Children's Hospital of Eastern Ontario and the Winnipeg Children's Hospital since 1985. Their surgical records, pathology reports, medical charts and radiographs before and after surgical excision were reviewed.

At surgery the protruding intra-articu-



FIG. 1. Anteroposterior (left radiograph) and lateral (right) views of patient A at 6 years of age, showing the typical medial involvement of the distal tibial epiphysis and medial talus. Note the misshapen contour of the medial part of the ankle.

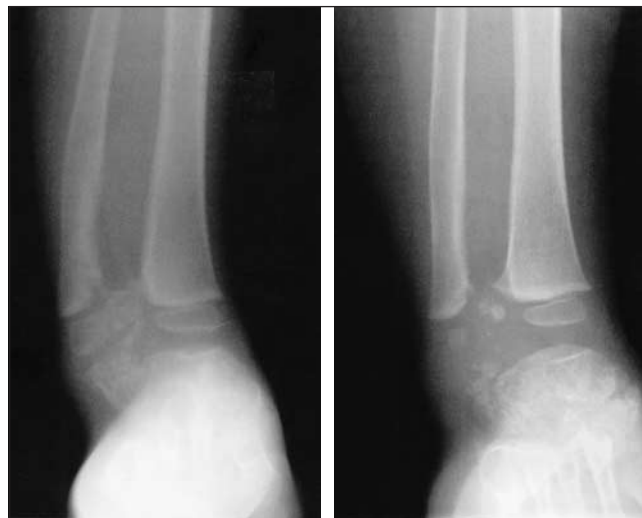


FIG. 2. Left, an anteroposterior view illustrating exostosis from the fibula epiphysis (patient B, at age 1½ yr). Right, a resection 1 year postoperatively, with the exostosis removed and showing a more normal-appearing ankle joint.

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lar cartilaginous mass was excised to restore joint congruity of the talus with the tibia and fibula, as well as to re-create the normal anatomy of the talus.

Results

All 4 children were boys, aged 1 to 4 years at presentation. Three were followed for an average of 3 years before surgical removal of the lesion. The other, with a lesion involving the fibula, underwent surgery shortly after diagnosis. Three required re-excision 5–7 years later. The longest follow-up was 11 years (patient C). Patient D is currently being followed subsequent to excision of a talar exostosis. The average follow-up from the first surgical treatment was 5.2 years; from diagnosis, 8.0 years. To date, none have required further surgery; patients A, B and C have exceeded skeletal maturity with no evidence of further growth of the lesions.

All had symptoms of pain in the ankle associated with running or long walks. Upon initial examination, all had restriction of dorsi-flexion and plantar flexion of the ankle of 10° or more compared with the contralateral side. Both ankle

pain and loss of motion increased with time, correlating with increasing joint incongruity as the lesion grew. At the initial surgery, all children were noted to have “misshapen ankles” (Fig. 1). Surgical excision of the DEH lesion resulted in improved plantar and dorsiflexion in all children at each surgical intervention (Fig. 2) to a normal range of passive motion comparable to the contralateral normal ankle. All children returned to full unrestricted activity after surgery. The 3 boys who required a second procedure to restore joint congruity began experiencing a recurrence of pain and limitation of ankle motion 3–4 years after the initial surgery.

All 3 children resumed their full physical education programs at school within 1 year of their second surgery and developed no later symptoms requiring further surgical intervention.

Discussion

Dysplasia epiphysealis hemimelica is a rare, unusual variant of osteochondroma primarily affecting the medial aspect of the epiphysis of the talus.

The 4 children in this review were all

severely compromised by DEH affecting the ankle resulting in joint incongruity, limitation of ankle motion and pain after lengthy walking or running. Three of these children were followed for 3 years with attempted activity modifications and ankle supports, which were unsuccessful due to the relentless and progressive growth of lesions (Fig. 3).

Surgical removal of the lesions provided considerable subjective and functional improvement in ankle function. Based on this review of 4 children, early removal of DEH lesions is recommended even though complete extirpation of the lesion is usually impossible without excising a segment of the talus or epiphysis large enough to result in permanent compromise of the ankle joint. The objective of the surgery should be to remove the joint incongruity caused by DEH to facilitate smooth function of the articulation, with the realization that the procedure may have to be repeated once or twice before skeletal maturity, when the continued growth of DEH appears to cease. Although 3 of these cases predated the availability of magnetic resonance imaging, imaging the ankle annually to ascertain the extent of cartilage growth and asymmetry secondary to DEH is recommended, so that the developing incongruity can be surgically corrected before secondary adaptive changes occur in the joint.

Conclusions

Dysplasia hemimelica epiphysealis, although rare, causes considerable disability in ankle function due to joint incongruity caused by the lesion's relentless growth during childhood. Early surgical removal to improve joint incongruity is recommended, with subsequent surgery to ensure continued congruity as required. Complete removal is usually not achievable initially. Children should be followed yearly, both clinically and with MRI, until the lesion stops growing at skeletal maturity.

Competing interests: None declared.

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FIG. 3. Left, anteroposterior radiograph of the ankle of patient C at presentation (age 3½ yr). The abnormal articulation of the right fibula and talus is obvious, with the foot being pushed into varus by the fibular epiphyseal expansion. Right, an anteroposterior view of the ankle taken 4 years later at age 7½. Note the continued enlargement of the fibular epiphysis and the intra-articular exostosis from the talus.

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Open tracheostomy in a suspect severe acute respiratory syndrome (SARS) patient: brief technical communication

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Patient management in the setting of severe acute respiratory syndrome (SARS) is complicated by the controversies about transmissibility, a reliable diagnostic tool and a clinically proven cure.¹⁻⁴ High-risk procedures are particularly problematic. Directives for high-risk procedures were published by Ontario's Ministry of Health and Long-Term Care in June 2003.⁵

Herein we describe the salient technical and essential infection-control principles⁶ (S. Abrahamson, unpublished data) learned from our experience with open tracheostomy in a SARS patient. We could find only 1 other report⁷ of tracheostomy in patients with or suspected of having SARS.

High-risk procedures such as intubation, bronchoscopy and tracheostomy should be done in a negative-pressure isolation environment. However, the Canadian Standards Association requires that operating rooms (ORs) operate at positive pressure. At present, there are no

negative-pressure isolation rooms in Toronto. A patient's clinical status or anatomy may warrant the greater anesthetic and surgical safety provided by ORs. If the patient's clinical status is expected to improve, it is prudent to wait; but difficult surgical anatomy is a more daunting problem, and should be given due consideration.

The possibility of contamination in the OR of health care workers and other patients should be balanced against the potential for surgical mishap in the intensive care unit (ICU). If OR personnel are educated in and equipped with personal protective systems, and the procedure planned in advance in consultation with the hospital's infection prevention and control service, the risk to health care workers is likely to be small.

Case

While driving, a 73-year-old man with ankylosing spondylitis was struck from

behind by another vehicle. He was brought to hospital 7 days after this motor-vehicle accident (MVA) with right torticollis and progressive numbness and weakness. A fracture/dislocation through C7-T1 was diagnosed, and he was transferred to St. Michael's Hospital for definitive management. Physical examination revealed profound quadriplegia, worse in the lower extremities and on the right. He experienced no bowel or bladder incontinence. Computed tomography confirmed angulation at C7/T1 with narrowing of his spinal canal; magnetic resonance imaging showed spinal-cord compromise.

The patient was managed with skeletal and halo traction, with only minimal improvement. On day 16 post-MVA, he underwent a posterior decompression, instrumentation and fusion from C3 to T3. The procedure was uncomplicated, and he was transferred to the trauma and neurosurgery ICU in stable condition.

His postoperative course was com-

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